Author's response to reviews

**Title:** Efficacy of aerobic physical retraining in a case of Combined Pulmonary Fibrosis and Emphysema Syndrome: a case report

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Efficacy of aerobic physical retraining in a case of Combined Pulmonary Fibrosis and Emphysema Syndrome: a case report

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Abstract

Introduction Combined Pulmonary Fibrosis and Emphysema (CPFE) has recently been recognized as a syndrome but remains under-diagnosed. Both clinical management and therapeutic approaches have not been clearly defined. Pulmonary rehabilitation has not been contemplated within therapeutic options for CPFE. In this case we explored the potential benefits of a specific aerobic physical retraining program in CPFE management.

Case presentation We describe a case of a 65 year old male caucasian affected by CPFE and respiratory failure receiving long term oxygen therapy (OTLT); the patient underwent physical retraining with moderate aerobic and breathing exercises for 4 weeks. Clinical and motor tests, as well as questionnaires assessing quality of life and depression levels, were performed prior to and following the retraining. At the end of retraining program a relevant reduction of long term oxygen therapy (OTLT) requirement was registered; improvements in terms of physical performance, quality of life and mood were observed in our patient but no change in respiratory parameters.

Conclusion A program of aerobic physical retraining would be beneficial in patients suffering from CPFE and should be considered as an additional therapeutic option.

Keywords

Combined Pulmonary Fibrosis and Emphysema, CPFE, Pulmonary rehabilitation, Aerobic physical retraining
Introduction
High-resolution CT (HRCT) scanning has enhanced recognition of the simultaneous occurrence of emphysema and pulmonary fibrosis and, recently, CPFE has been defined as a syndrome. This syndrome is characterized by upper lobe emphysema and lower lobe fibrosis (1).
Common risk factors resulting in epithelial lung alterations may contribute to emphysema and pulmonary fibrosis despite clinical, radiological and pathologic differences (2-4). Coexistence of emphysema and fibrosis are more likely to occur in smoking-induced parenchymal pulmonary damage (5-6). CPFE is still under-diagnosed and clinical management as well as therapeutic approaches are object of debate. In this case we explore the potential benefits of a specific aerobic physical retraining program which has not yet been considered within therapeutic options for CPFE.

Case presentation
We report a case of a 65 year old male, Caucasian, ex-smoker (40 pack-years) affected by CPFE with several comorbidities including hypertension, type II diabetes and depression. Before CPFE diagnosis the patients has been affected by chronic bronchitis for 12 years. Diagnosis of CPFE was confirmed by thoracic HRCT. On admission the patient, exhibited respiratory failure, reporting dyspnea on exertion, productive cough; the patient received long term oxygen therapy (LTOT) with 2,5 L/min of flow during 24 hours. Spirometry showed relatively a mild restriction although Diffusion Lung Capacity for Carbon Monoxide (DLCO) was 44% of predicted. Concurrent pulmonary hypertension (PAPs 60 mmHg) was diagnosed by color-doppler echocardiography. Pharmacological therapy consisted of high dose inhaled steroids.
Following clinical, respiratory and physical exercise test assessments, quality of life (St. George Respiratory) and depression level (Geriatric Depression Scale) questionnaires were administered. The patient was then enrolled in the retraining program for four weeks. Written consent was obtained from the patient before starting the exercise program.
Each retraining session was supervised by an exercise physiologist monitoring heart and respiratory rate, oxygen saturation, blood pressure and level of dyspnea. The patient underwent the exercise program for 2 sessions per day up to 30 minutes each, 5 days per week, for a 4 week period. The training included one session of aerobic exercise and one session of breathing technique. For aerobic training, the subject exercised on a treadmill, with a speed of 2.5 km per hour and 0% of slope. The work rate of training corresponded to 50%-60% of the maximum heart rate and the training began at a work rate equal to 50%. When the patient reached the level of exercise at 10 minutes without intolerable dyspnea (Borg rating of breathlessness of < 5), the workload (speed and/or elevation) and duration of exercise were increased by 10% (7).

Progressive exercise training was employed for respiratory muscles; the training consisted of diaphragmatic breathing and inspiratory muscle training, through an inspiratory threshold, at 40% - 50% of the initial Pimax. During training the patient received LTOT with a flow reduction of 15% each week.

After the 4 weeks retraining program LTOT required to correct hypoxemia was decreased to a flow of 1.5 L/min for 24 hours; arterial blood gas analysis demonstrated a PaO2 increase of 11% and PCO2 decrease of 5.6%. The reason for the oxygen requirement reduction observed was probably due to the strengthening of respiratory muscles which allowed a better oxygenation of skeletal muscles as a result of both aerobic and breathing exercises (8). Estimated pulmonary artery systolic pressure by trans-thoracic two-dimensional echocardiography improved by 16 % in terms of cardiac index and pulmonary vascular resistance (9); no significant lung function changes were observed. Relevant improvements in exercise capacity, dyspnea rating, health-related quality of life and levels of depression were also demonstrated after the pulmonary rehabilitation program.

The 6'WT showed an increase of distance walked by 133 %. The dyspnea score was reduced by 1.0, 3.0 and 4.0 points with only a minor increased of Pimax of the 17%. Patient adherence to
retraining programme was good. Table1 reports the test results collected before and after the retraining program.

Discussion

In this case study we have evaluated whether a patient with CPFE, a disease which has not yet been considered for pulmonary rehabilitation, can benefit from an aerobic retraining program. Enphysema and Pulmonary Fibrosis are progressive lung diseases which may be associated with co-morbidities and systemic consequences (10-14). CPFE syndrome typically occurs in male smokers and is frequently complicated by pulmonary hypertension, acute lung injury, and lung cancer which affects disease progression and survival.

In the literature there are no reports on the efficacy of physical retraining in CPFE patients, in contrast with other lung diseases such as COPD or IPF where clinical studies have demonstrated the benefits of pulmonary rehabilitation (15).

Clinically relevant improvements and short term benefits were clearly demonstrated in this report as a result of a retraining program. Although clinical studies on long term effects within a large patient population are required for definitive conclusions our observations suggest that integrating pulmonary rehabilitation with pharmacological treatments would be beneficial for CPFE patient.

Conclusion

In the literature there are no reports on the effects of pulmonary rehabilitation on CPFE patients. The efficacy of aerobic exercise on quality of life and psychological well-being and exercise capacity function on CPFE patient has been demonstrated in this case. Our results suggest that exercise should be considered as a therapeutic strategy for CPFE patients, although further studies considering the impact of different types of training in these subjects are required.
Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

FEV1, Forced Expiratory Volume in one second; FVC, Forced Vital Capacity; PaO2, Partial Arterial Pressure of Oxygen; PaCO2, Partial Pressure of Carbon Dioxide; PAP, Pulmonary Artery Pressure; PiMax, Maximal Inspiratory Pressure; PeMax, Maximal Expiratory Pressure

Competing interests

The authors declare that they have no competing interests.

Authors’ contributions

GDS, GM, AB, GC analysed and interpreted the patient data and wrote the article, GDS, AG, CDG, planned and carried out the pulmonary rehabilitation program. All authors read and approved the final manuscript.

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